NOX2 in the hSOD1G93A Transgenic Swine: a preliminary overview

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Since 1993 some forms of amyotrophic lateral sclerosis (ALS) have been associated with several mutation of superoxide dismutase 1 (SOD1) gene. A number of ALS animal models have therefore been developed based around this gene, although with doubts about their suitability to reproduce human disease.
Recently, a transgenic swine ALS model expressing mutant human SOD1^{G93A} (glycine 93 changed to alanine) was generated by in vitro transfection of cultured somatic cells and used as new and alternative model to study ALS neuropathology due to closer anatomical, physiological and biochemical features of pig rather than rodents to humans.
In this study, the same NADPH oxidase-2 (NOX2) and SOD1 analyses performed in our ALS patients were carried out on blood aliquots of two transgenic swine and one control.
The goal of this part of Neurinox project was to compare findings of SOD1^{G93A} pig with the human mutations G93D, A4V, L144F, T137A, I113F and N19s, in order to investigate similarities and differences and verify the suitability of pig model to reproduce the ALS of SOD1 mutated humans.

Reference
Modeling amyotrophic lateral sclerosis in hSOD1 transgenic swine
Chieppa MN et al. Neurodegener Dis. 2014;13(4):246-54.